

The Texas Birth Defects MONITOR

An Annual Data and Research Update



Volume 22

December 2016

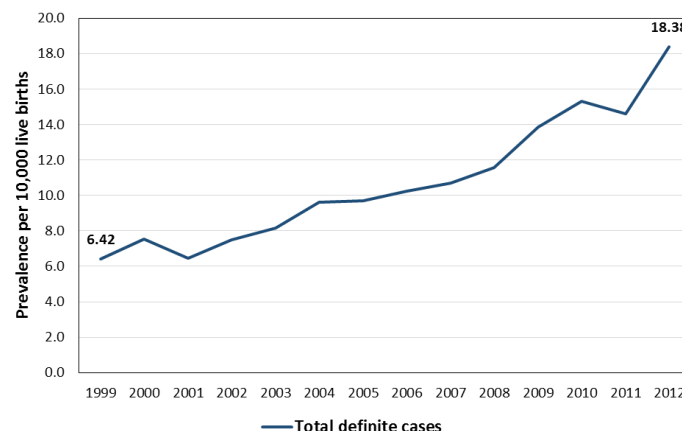
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Highlight on Microcephaly

Recently a growing interest in microcephaly emerged due to the association of the condition with the Zika virus. Microcephaly is diagnosed when a baby's head is smaller than expected compared to babies of the same gestational age and sex.

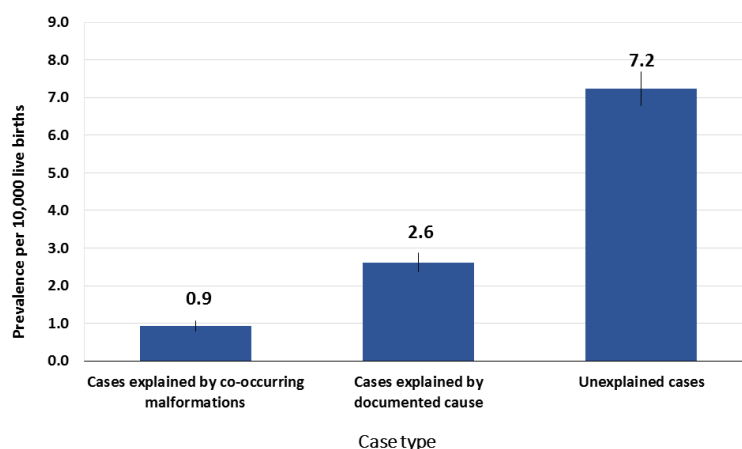
Highlight on Microcephaly	1	Several years ago, the Texas Birth Defects Registry stopped routinely publishing data on microcephaly because of the broad case definition that Texas uses for surveillance of the condition. However, the Registry continued to collect data on microcephaly, as well as on conditions that occur with microcephaly. Staff recently conducted numerous epidemiologic analyses of microcephaly, prior to the arrival of Zika in Texas.
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Updated Texas Birth Defects Registry Data	4	The chart below demonstrates a threefold increase in the birth prevalence of definite cases of microcephaly over a 15-year period. Cases were included regardless of the cause of the defect or its severity. It is important to note, however, that about 30% of all cases are explained by a co-occurring condition (e.g., other brain/skull defects) or a documented cause (e.g., chromosomal anomalies, infections, alcohol exposure). The prevalence of definite microcephaly by subgroup is shown in the chart on page two.
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Microcephaly: Total Definite Cases, Texas, 1999-2012



(Highlight on Microcephaly, continued from page 1)

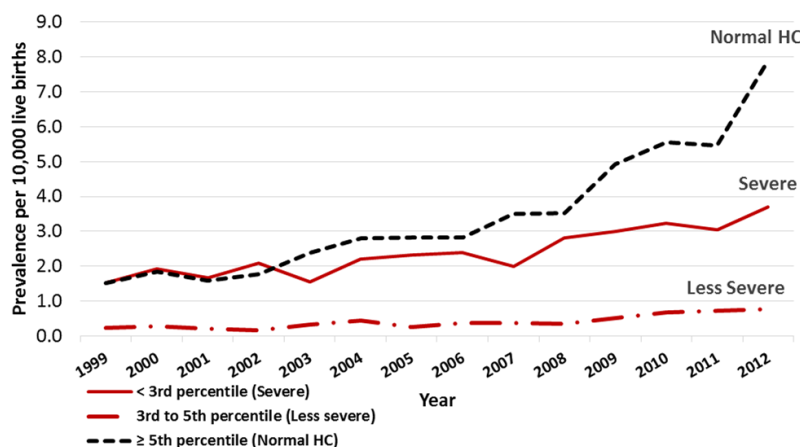
Prevalence of Definite Microcephaly, by Subgroup, Texas, 1999-2012



Microcephaly can have varying levels of severity, depending in part on how small the head is relative to infants of the same sex and gestational age. Thus, the Registry determines the severity of microcephaly by comparing the head circumference of the newborn collected at delivery against growth charts recommended by the National Birth Defects Prevention Network and the Centers for Disease Control and Prevention. Babies born with a head circumference less than the 3rd percentile for infants of the same gestational age and sex are considered to have “severe microcephaly.” Severe unexplained microcephaly made up 21% of all cases. Severe unexplained microcephaly was more common among non-Hispanic black mothers and female infants.

The time trend described above is driven by a substantial, fivefold, increase in unexplained microcephaly cases with a normal head circumference at delivery (greater than the 5th percentile) as depicted in the figure below. The increasing time trend can be a consequence of changes over time in the diagnosis, recording, and ascertainment of microcephaly. On the other hand, the prevalence of severe unexplained microcephaly cases increased by only twofold.

Unexplained Cases of Definite Microcephaly, by Percentile of Head Circumference at Delivery, Texas, 1999-2012



January is National Birth Defects Prevention Month

Prevent to Protect: Prevent Infections for Baby's Protection

Birth defects are common, costly, and critical. Every 4½ minutes, a baby is born with a major birth defect in the United States. Become an active participant in National Birth Defects Prevention Month by joining the nationwide effort to raise awareness of birth defects, their causes, and their impact.

Infections during pregnancy can hurt both mothers and their babies. We encourage all women to make healthy choices and learn different strategies to prevent infections during pregnancy to help lower their risk of having a baby born with a birth defect. This year we are encouraging all women to prevent infections to protect their babies by observing the following guidelines:

- **Properly prepare food.**
 - ◇ Wash your hands before and after preparing food.
 - ◇ Do not eat raw or runny eggs or raw sprouts.
 - ◇ Avoid unpasteurized (raw) milk and cheese, and other foods made from them.
- **Talk to your healthcare provider.**
 - ◇ Talk to your healthcare provider about what you can do to prevent infections such as Zika virus.
 - ◇ Make sure that you are up-to-date with vaccinations (shots) before getting pregnant.
 - ◇ Talk to your healthcare provider about vaccinations that you should receive during pregnancy.
- **Protect yourself from animals and insects known to carry diseases such as Zika virus.**
 - ◇ Stay away from wild or pet rodents, live poultry, lizards and turtles, and do not clean cat litter boxes while pregnant.
 - ◇ When mosquitoes and ticks are active, wear long-sleeved shirts and long pants when outside.
 - ◇ Use Environmental Protection Agency (EPA) registered insect repellents with one of the following active ingredients: DEET, picaridin, IR3535, or oil of lemon eucalyptus (para-menthane-3,8-diol).
- **Maintain good hygiene.**
 - ◇ Wash your hands often with soap and water, especially
 - Before preparing or eating foods,
 - After handling raw meat, raw eggs, or unwashed vegetables,
 - After being around or touching pets and other animals,
 - After changing diapers or wiping runny noses.
 - ◇ Do not put a young child's food, utensils, drinking cups, or pacifiers in your mouth.

Women and their loved ones can participate in these strategies and take these important steps toward a healthy pregnancy. Share your own tips for a healthy pregnancy using #Prevent2Protect on social media. Learn more at www.nbdpn.org.

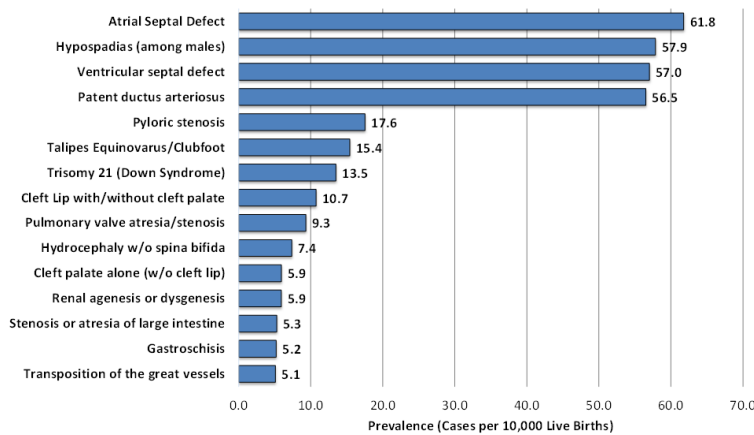
Source acknowledgement: National Birth Defects Prevention Network Education and Outreach Committee.

Updated Texas Birth Defects Registry Data, 1999-2013

The Texas Birth Defects Registry has released its Report of Birth Defects Among 1999-2013 Deliveries. The full report shows the birth prevalence of selected defects among deliveries to Texas residents during 1999-2013 by delivery year, mother's age, mother's race/ethnicity, sex of the infant or fetus, and region of the state, as well as by pregnancy outcome.

The chart below illustrates the 15 most common birth defects in Texas from 1999-2013. The most common birth defect was atrial septal defect, a heart defect with a prevalence of about 62 cases per 10,000 live births (0.6%, or one in every 169 births).

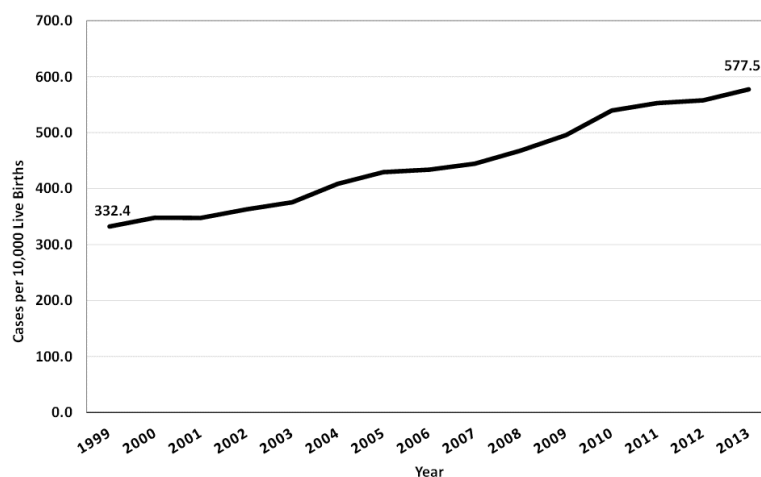
The 15 Most Common Birth Defects in Texas, 1999-2013*



**Among the birth defects that are routinely published by the Texas Birth Defects Registry*

The overall prevalence of birth defects is increasing with time, as shown in the figure below. We believe that most of the observed increasing trend is explained to an increase in the awareness, documentation, and detection of birth defects during infancy (Langlois et al., Birth Defects Res A Clin Mol Teratol 2011). Not all birth defects show an increasing trend with time. The prevalence of births defects may decrease, increase, or remain about the same from year to year.

Prevalence of Any Monitored Birth Defect Over Time, Texas, 1999-2013

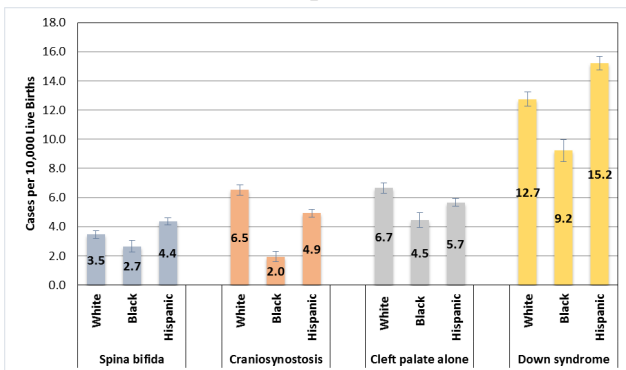


Maternal Race/Ethnicity and Birth Defects in Texas, 1999-2013

There are numerous differences in the prevalence of birth defects across maternal racial/ethnic groups. Overall, Non-Hispanic (NH) White mothers are 7% more likely than Non-Hispanic (NH) Black and Hispanic mothers to have a baby with any monitored birth defect.

The figure below shows the prevalence of spina bifida, craniosynostosis, cleft palate alone, and Down syndrome among three racial/ethnic groups (NH White, NH Black, and Hispanic). The graph shows the varying prevalence of these conditions according to racial/ethnic group. For each defect depicted, all racial/ethnic prevalences are significantly different from each other.

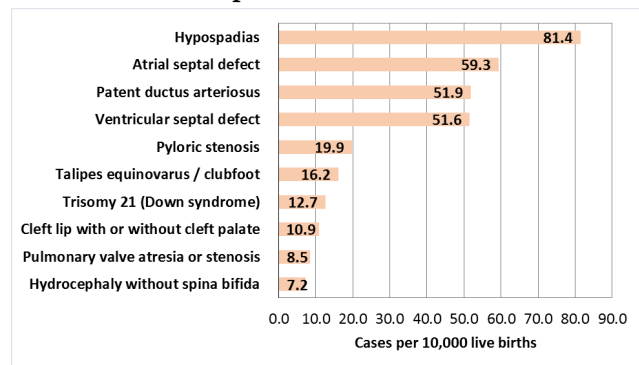
Selected Birth Defects with Statistically Significant Differences in Prevalence among Three Racial/Ethnic Groups, Texas, 1999-2013*



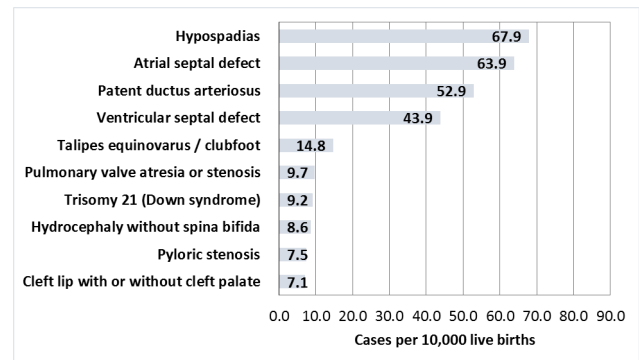
For instance, craniosynostosis and cleft palate alone are significantly more prevalent among infants of NH White mothers, compared to infants of NH Black and Hispanic mothers. For spina bifida and Down syndrome, the prevalence is higher among infants of Hispanic mothers, compared to infants of NH White and NH Black mothers.

To further show how the prevalence of birth defects can vary based maternal race/ethnicity, the figures below show the 10 most common birth defects among infants of NH White, NH Black, and Hispanic mothers, ranked by prevalence.

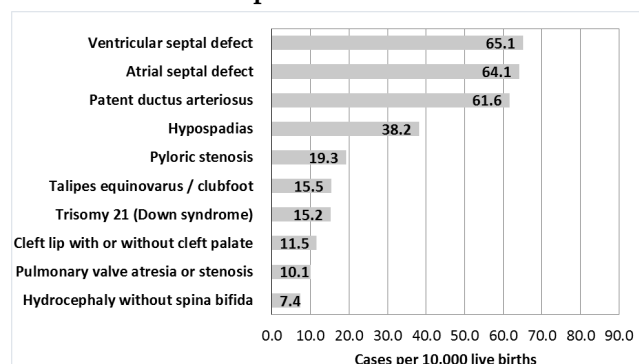
10 Leading Birth Defects in Infants of Non-Hispanic White Mothers*



10 Leading Birth Defects in Infants of Non-Hispanic Black Mothers*



10 Leading Birth Defects in Infants of Hispanic Mothers*



*Among the birth defects that are routinely published by the Texas Birth Defects Registry

(Continued on page 6)

The three leading birth defects among infants of NH White and NH Black mothers are the same: hypospadias, atrial septal defect, and patent ductus arteriosus (shown in the first two bar charts on the previous page). However, the three leading birth defects of infants born to Hispanic mothers are different from the other two groups: ventricular septal defect, atrial septal defects, and patent ductus arteriosus (shown in the third bar chart on the previous page). Prevalence of pyloric stenosis was significantly higher in infants of NH White and Hispanic mothers (19.9 and 19.3 cases per 10,000 live births respectively), compared to infants of NH Black mothers (7.5 cases per 10,000 live births).

Texas Birth Defects Epidemiology and Surveillance Branch Receives New Grant Funding through CDC

The Texas Birth Defects Epidemiology and Surveillance Branch has received two new grants (Cooperative Agreements) from the Centers for Disease Control and Prevention (CDC).

The first, "Population-based Surveillance of Birth Defects and Data Utilization for Public Health Action," aims to improve the way the BDES branch utilizes its data. The purposes of the grant are to evaluate the program's case ascertainment and enhance surveillance; expand the program's reach (with new partners and stakeholders); and conduct analytical activities related to critical congenital heart defects. The grant funds two new positions within the branch.

The other cooperative agreement, "Surveillance, Intervention, and Referral to Services Activities for Infants With Microcephaly," seeks to establish, enhance and maintain rapid population-based surveillance of microcephaly and related conditions (especially central nervous system defects) possibly linked to Zika virus infection during pregnancy. In addition, the grant allows for participation in data projects, referral of affected infants and families to services, and assessing outcomes of affected children. The grant funds eight new positions, mostly related to data collection in regional offices.

Save the Date

The 44th Annual Meeting of the International Clearinghouse for Birth Defects Surveillance and Research will be held November 13-15 in Austin, Texas at the Sheraton Austin Hotel at the Capitol.

Sign up for GovDelivery email updates from the Texas Birth Defects Epidemiology and Surveillance Branch on the BDES website at <https://www.dshs.texas.gov/birthdefects/>.

Associations between Maternal Periconceptional Exposure to Secondhand Smoke and Major Birth Defects

The relationship between exposure to secondhand smoke (SHS) during pregnancy and birth defects needs further research and examination. SHS remains an important public health concern, especially in groups where exposure appears to be higher. This study used data from the multisite case-control National Birth Defects Prevention Study (NBDPS) to determine whether mothers with any SHS exposure during pregnancy gave birth to more infants with birth defects compared to mothers with no SHS exposure during pregnancy.

This study examined the association between maternal periconceptional exposure to secondhand smoke (SHS) and a spectrum of major birth defects for delivery years 1997 through 2009. The analysis was restricted to birth defects with ≥ 100 cases, singleton births, non-smokers, and mothers without pregestational type- 1 or 2 diabetes (which has been associated with a range of birth defects). Following the inclusion/exclusion criteria, 44 birth defect groups were identified –leaving a total of 18,762 cases and 7,747 controls. Maternal exposure to SHS was examined for the period one month prior to conception through the first trimester for all birth defects; with the exception being craniosynostosis—where associations in the second and third trimesters were also examined. Additionally, source of SHS exposure (whether household or workplace/school – occurring concurrently or independently) were also examined. Multivariate logistic regression models were used to estimate both crude and adjusted odds ratios and 95% confidence intervals (cORs/aORs, 95% C.I.s). The odds ratios were adjusted for a variety of maternal and infant factors including: maternal age, education, race/ethnicity, BMI, periconceptional

alcohol and folic acid intake, previous live births, maternal nativity, pregnancy intention, household income/number of people supported by the income, study center, and time to interview.

The authors found that the prevalence range of SHS was wider for cases (12.9-27.8%) than for controls (14.5-15.8%). A modest positive association between exposure to any SHS and 8 birth defects, 1 Congenital Heart Disease (CHD) and 7 non-CHDs, was observed. Positive associations were noted between SHS and the following groups:

- Neural tube defects (anencephaly: (aOR, 1.66; 95% C.I., 1.22-2.25); spina bifida: (aOR, 1.49; 95% C.I., 1.20–1.86)),
- Orofacial clefts (cleft lip without cleft palate: (aOR, 1.41; 95% C.I., 1.10-1.81); cleft lip with or without cleft palate (aOR, 1.24; 95% C.I., 1.05-1.46); and cleft palate alone (aOR, 1.31; 95% C.I., 1.06-1.63)),
- Bilateral renal agenesis (aOR, 1.99; 95% C.I., 1.05-3.75), amniotic band syndrome-limb body wall complex (aOR, 1.66; 95% C.I., 1.10-2.51),
- Atrial septal defects, secundum (aOR, 1.37; 95% C.I., 1.09-1.72).

No significant inverse associations were observed across any of the case groups.

The results are consistent with previous studies which have found SHS to be moderately associated with a few birth defects—particularly NTDs and orofacial clefts. Despite the moderate size of the risk estimates, the pervasiveness of SHS worldwide, particularly in lower-resource countries and segments of the population, may translate into many cases of potentially preventable birth defects.

Information about Down Syndrome for New and Expecting Parents

A new brochure with information on Down syndrome for new and expecting parents is now available through the Texas Department of State Health Services website. This brochure, available in English and Spanish, contains important information on the condition and resources for families. The brochure can be found here: www.dshs.texas.gov/birthdefects/downsyndrome.

This brochure was a product of the implementation of House Bill 3374, which became effective in 2015. This legislation requires Texas health care providers to provide information to expectant or new parents following their child's diagnosis of Down syndrome.

Below are some recommended resources for new and expectant parents of a child with the condition. These materials have been reviewed by medical experts and are included in the "National Society of Genetic Counselors practice guidelines for communicating a prenatal or postnatal diagnosis of Down syndrome."

- **"Diagnosis to Delivery: A Pregnant Mother's Guide to Down Syndrome"** By Nancy McCrea Iannone and Stephanie Hall Meredith, Down Syndrome Pregnancy, www.downsyndromepregnancy.org/books/. A book and website with information for new and expectant parents anticipating the birth of a child with Down syndrome.
- **"Facts about Down Syndrome"** Centers for Disease Control and Prevention (CDC), www.cdc.gov/ncbddd/birthdefects/DownSyndrome.html. A website with facts about Down syndrome.
- **"Babies with Down Syndrome"** Edited by Susan J. Skallerup, Woodbine House, Books on Disabilities. An extensive book for new parents of a child with Down syndrome. ("Bebés con síndrome de Down" — Spanish edition.)
- **"Gifts: Mothers Reflect on How Children with Down Syndrome Enrich Their Lives"** Edited by Kathryn Lynard Soper, Woodbine House, Books on Disabilities. Mothers' stories describing the gifts that children with Down syndrome have brought into their lives.
- **"What Parents Wish They'd Known: Reflections on Parenting a Child with Down Syndrome"** Edited by Kathryn Soper, Woodbine House, Books on Disabilities. Book with personal essays and quotes by new and expectant parents of a child with Down syndrome.
- **"Health Care Information for Families of Children with Down Syndrome"** American Academy of Pediatrics, www.healthychildren.org. Guidelines focused on recommended healthcare and treatment of a child with Down syndrome.
- **"Planning for a Loving Future"** National Down Syndrome Adoption Network, www.ndsan.org/about-ndsan/. Pamphlets and website for parents considering adopting a child with Down syndrome.
- **"A Loving Choice: Making an Adoption Plan for a Child with Down Syndrome"** National Down Syndrome Adoption Network, www.ndsan.org/adoption-process/birth-parents/. Pamphlets and website for parents considering placing their child for adoption.

For more information and resources, visit www.dshs.texas.gov/birthdefects/downsyndrome.

Folic Acid Awareness Week

January 8-14, 2017

January 8-14 is Folic Acid Awareness Week, but folic acid should be part of a healthy lifestyle every day.

What is folic acid and why do you need it?

- Folic acid is an essential B-vitamin; therefore, everyone needs it in order to stay in good health. Folic acid helps build DNA and your body uses it for cell growth and reproduction, fundamental building block processing and genetic material production.
- In 1998, the U.S. Food and Drug Administration started fortifying cereal grain products with folic acid in order to reduce the risk for neural tube defects (NTDs), serious birth defects of the brain and spine. While this was a great step in the fight to prevent birth defects, it is often not enough to protect all women and their potential children.
- Folic acid is water soluble, therefore it passes through your body very quickly. Taking folic acid every day ensures that you always have it in your system when your body needs it.
- It is particularly important for women of reproductive age to get 400 mcg of folic acid daily. It has been shown to reduce the risk of having an NTD by up to 70 percent. Since about 50 percent of pregnancies in the United States are unplanned, it's important to take folic acid every day, even if you're not planning to get pregnant. The most common NTDs are spina bifida and anencephaly.

Who needs folic acid?

All women need folic acid every day. Getting enough folic acid every day, before and during early pregnancy, is an important way to reduce the risk of NTDs. These birth defects occur in the first weeks of fetal development, often before a woman even knows she is pregnant.

Source acknowledgement: National Birth Defects Prevention Network Education and Outreach Committee.



The International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR) and participating organizations are raising awareness of birth defects with World Birth Defects Day, held on March 3. The ICBDSR hopes to increase global awareness of birth defects and promote expansion of birth defects surveillance, prevention, research, and care with this event.

To learn more, visit <http://www.icbdsr.org/>.

Contribution of Birth Defects to the Leading Causes of Death by Age Group, Texas, 2013*

The table below lists the leading causes of death by age group in Texas for 2013. Birth defects are the leading cause of infant mortality in Texas, and also remain a significant cause of death among Texans into young adulthood.

	<1 year of age	1-4 years of age	5-14 years of age	15-24 years of age
Rank				
1	Birth Defects 525 deaths	Unintentional Injuries 148 deaths	Unintentional Injuries 141 deaths	Unintentional Injuries 1,134 deaths
2	Short Gestation/Low Birth Weight 340 deaths	Malignant Neoplasms 49 deaths	Malignant Neoplasms 109 deaths	Suicide 419 deaths
3	SIDS 153 deaths	Birth Defects 43 deaths	Birth Defects 33 deaths	Homicide 310 deaths
4	Maternal Pregnancy Complications 145 deaths	Homicide 28 deaths	Suicide 27 deaths	Malignant Neoplasms 141 deaths
5	Unintentional Injury 83 deaths	Heart Disease 22 deaths	Chronic Lower Respiratory Disease 20 deaths	Heart Disease 72 deaths
6	Placenta, Cord, and Membrane Complications 80 deaths	Certain Conditions Originating in the Perinatal Period 9 deaths	Homicide 19 deaths	Birth Defects 29 deaths
7	Bacterial Sepsis 55 deaths	Chronic Lower Respiratory Disease 9 deaths	Heart Disease 14 deaths	Cerebrovascular Diseases 20 deaths
8	Respiratory Distress of Newborn 53 deaths	Influenza/Pneumonia 8 deaths	Cerebrovascular Diseases 12 deaths	Septicemia 18 deaths
9	Neonatal Hemorrhage 51 deaths	Septicemia 4 deaths	Influenza/Pneumonia 10 deaths	HIV 18 deaths
10	Assault (Homicide) 42 deaths	3 conditions 3 deaths each	Septicemia 5 deaths	Influenza/Pneumonia 18 deaths

*Data taken from the 2013 Vital Statistics Annual Report, Center for Health Statistics, Texas Department of State Health Services.

Recent Publications from BDES Branch Staff and Collaborators

Agopian AJ, Hoang TT, Mitchell LE, Morrison AC, Tu D, Nassar N, Canfield MA. Maternal hypertension and risk for hypospadias in offspring. *Am J Med Genet A*. 2016 Aug 29. doi: 10.1002/ajmg.a.37947. [Epub ahead of print]

Gong X, Zhan FB, Brender JD, Langlois PH, Lin Y. Validity of the emission weighted proximity model in estimating air pollution exposure intensities in large geographic areas. *Sci Total Environ*. 2016 May 3; 563: 478-485.

Hoyt AT, Canfield MA, Romitti PA, Botto LD, Anderka MT, Krikov SV, Tarpey MK, Feldkamp ML. Associations between Maternal Periconceptional Exposure to Secondhand Tobacco Smoke and Major Birth Defects. *Am J Obstet Gynecol*. 2016 Jul 18. pii: S0002-9378(16)30456-2. doi: 10.1016/j.ajog.2016.07.022. [Epub ahead of print]

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Kim J, Langlois PH, Herdt-Losavio ML, Agopian AJ. A Case-Control Study of Maternal Occupation and the Risk of Orofacial Clefts. *J Occup Environ Med*. 2016 Jul 12. [Epub ahead of print] PMID: 27414011 DOI: 10.1097/JOM.0000000000000807

Lara DA, Ethen MK, Canfield MA, Nembhard WN, Morris SA. A population-based analysis of mortality in patients with Turner syndrome and hypoplastic left heart syndrome using the Texas Birth Defects Registry. *Congenit Heart Dis*. 2016 Sep 29. doi: 10.1111/chd.12413 [Epub ahead of print].

Lara DA, Fixler DE, Ethen MK, Canfield MA, Nembhard WN, Morris SA. Prenatal diagnosis, hospital characteristics, and mortality in transposition of the great arteries. *Birth Defects Research Part A: Clinical and Molecular Teratology*. 2016 June 14. doi:10.1002/bdra.23525. [Epub ahead of print]

Lee LJ, MPH, Symanski E, Lupo PJ, Tinker SC, Razzaghi H, Pompeii LA, Hoyt AT, Canfield MA, Chan W, The National Birth Defects Prevention Study. Data linkage between the National Birth Defects Prevention Study and the Occupational Information Network (O*NET) to assess workplace physical activity, sedentary behaviors, and emotional stressors during pregnancy. *Am J Indus Med*. 2016;59:137-149.


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O'Brien JL, Langlois PH, Lawson CC, Scheuerle A, Rocheleau CM, Waters MA, Symanski E, Romitti PA, Agopian AJ, Lupo PJ; National Birth Defects Prevention Study. Maternal occupational exposure to polycyclic aromatic hydrocarbons and craniosynostosis among offspring in the National Birth Defects Prevention Study. *Birth Defects Res A Clin Mol Teratol*. 2016;106(1):55-60.

Vuong AM, Shinde MU, Brender JD, Shipp EM, Huber Jr. JC, Sharkey JR, McDonald TJ, Werler MM, Kelley KE, Griesenbeck JS, Langlois PH, Canfield MA, National Birth Defects Prevention Study Investigators. Prenatal Exposure to Nitrosatable Drugs, Dietary Intake of Nitrites, and Preterm Birth. *Am J Epi*. 2016; doi:10.1093/aje/kwv250.

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| 2017 Calendar

 The Monitor is published annually by the Birth Defects Epidemiology and Surveillance Branch, Texas Department of State Health Services.

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Please visit the BDES website for updated information and to sign up for GovDelivery updates: www.dshs.texas.gov/birthdefects/.

Requests for copies or back issues may be made to: birthdefects@dshs.texas.gov.

- January: National Birth Defects Prevention Month
- January 8-14: National Folic Acid Awareness Week
- February 14: Congenital Heart Defect Awareness Day
- January 23-28: Society for Maternal-Fetal Medicine Annual Meeting, Las Vegas, Nevada
- Spring 2017: March of Dimes March for Babies (check with MOD for specific dates and locations)
- March 3: World Birth Defects Day
- March 5-8: National Family Planning & Reproductive Health National Conference, Washington, DC
- April: National Autism Awareness Month
- April: Alcohol Awareness Month
- April 3-9: National Public Health Week
- June 18-20: Annual Meeting of the Society for Pediatric and Perinatal Epidemiologic Research, Seattle, WA
- June 24-28: 55th Annual Meeting of the Teratology Society, Denver, Colorado
- July: National Cleft and Craniofacial Awareness & Prevention Month
- July: Gastroschisis Awareness Month
- September: Childhood Cancer Awareness Month
- September: Newborn Screening Awareness Month
- October: National Spina Bifida Awareness Month
- October: National Down Syndrome Awareness Month
- October 2-4: International Conference on Neural Tube Defects, Austin, Texas
- November: Prematurity Awareness Month (March of Dimes)
- November 13-15: 44th Annual Meeting of the International Clearinghouse for Birth Defects Surveillance & Research, Austin, Texas